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Art. II.—Account of the POST-MORTEM examination of a Case of DEAFNESS, in which were found an abnormal State of the OSSICULA AUDITUS, with other irregularities in one Ear, and Destruction of the MEMBRANA TYMPANI and disorganization of the soft parts in the Tympanum, &c. in the other.

BY JOSHUA I. COHEN, M. D. OF BALTIMORE, MD.

[The following valuable communication was read before the American Philosophical Society, at a recent meeting; and is noticed in the "Proceedings" of that body. It has rarely happened that opportunities have been embraced for examining into the condition of the organ of hearing in cases of deafness, or that they have fallen within the observation of an investigator so competent as the author of this Paper.—Ed.]

The facts contained in the following paper have been deemed worthy of note, as adding one more example to the few on record of the undeveloped or rudimentary condition of a portion of the organ of hearing in man. For this reason the case has been thought of sufficient interest to claim the honour of the attention of the members of this society. Although the irregularity belongs to a very minute structure of the human body, it is nevertheless connected with an important organ, whose whole structure is minute, and in which every deviation, however slight, from the usual normal state, must influence, more or less, the proper exercise of its function.

Those members of the society who have been engaged in investigating the pathological conditions of the organ of hearing, and have felt, like myself, even with our present improved methods of investigation, the difficulties attendant thereupon, from the want of a series of well-observed facts, will I am sure, receive with due attention, every contribution that may tend, in the slightest degree, to illustrate the condition upon which manifestations of disease depend.

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Case.—While engaged in September last in some investigations upon the ear, at the Baltimore County Alms-house, my attention was called by Dr. Cunningham, one of the resident physicians, to the case of a man (æt. about 40) who had just died, and who, during his residence of six months in the house, laboured under great difficulty of hearing. He died of tubercular phthisis, and the examination which Dr. C. made, showed the usual condition of the lungs in that disease.

Glad of the opportunity so rarely afforded, to investigate *post-mortem* the condition of the organs of hearing of one who, it was known, was deaf, I readily availed myself of the information, and proceeded at once to examine the state of the brain and seventh pair of nerves. These parts were carefully dissected, but there was observed no change from the usual condition.

In order to examine at greater leisure, and more carefully than I could do at the moment, the several parts of the minute apparatus of the organs of hearing, and particularly to note the condition of the Eustachian tubes, a portion of their structure which has lately attracted much attention in cases of deafness, I removed the whole lower part of the skull, so as to preserve the connection of these tubes, and on the following day observed the appearances now to be described.

Eustachian Tubes.—These were found in nearly the same state on each side: their guttural orifices were unusually large. The cartilages around them were much developed, particularly that of the right tube. The tubes themselves were unobstructed in their whole extent, and their diameters even larger than common.

Right Ear.—The external ear and meatus presented the usual appearance—the latter having been sawed off so as to expose the membrana tympani; this membrane, instead of the uniform, clear, semi-transparent and glistening character usually observed, with its concavity only towards the middle, was of a dull and dark appearance, irregular and evidently thickened; the concavity extending from the circumference, saucer-like: the whole membrane being drawn inwards.

The bone was now cut through so as to show the cavity of the tympanum. This was found to be very much narrowed by the approximation of the membrana tympani to the opposite walls; and, instead of a clear, empty space for the ossicles, &c. the tympanum was filled with muco-fibrous membranes passing from the membrana tympani to the posterior wall, presenting a cellular structure.

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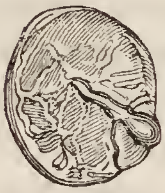
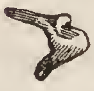
Perceiving some irregularity in the ossicula, I carefully divided these membranes, in order to get a distinct view of the bones *in sitû:* when the *tensor tympani muscle* was exposed, I found that the membrana tympani with its ring of bone was only bound down in its place by this connection; the chain of bones being incomplete. Before separating this bond of union, I noted the unusual shortness of the tendon of the muscle of the malleus, and that it was attached to the handle of this bone throughout its whole length, thus drawing down the bone, and with it the membrana tympani to within a line of the cochleariform process. This tendon having been divided, and the membrana tympani with its ring of bone separated, the following abnormal state of the bones was seen:—

Ossicula Auditûs.—The *malleus* was normal, but its relation to its proper muscle irregular, as above described.





Incus.—This bone was undeveloped. Its size and form will be better understood by a reference to the accompanying drawing. It has a small articulating surface, by which it is closely attached to the corresponding surface of the malleus—from this a very short process extends backwards towards the mastoid cells, not reaching, however, beyond the circumference of the membrana tympani, to which it was attached closely by a thick fold of membrane reflected from the latter. This constituted the whole bone.

Stapes.—The stapes was wanting, with the exception of the *base*—this was nicely adapted to the size of the fenestra ovalis, leaving, as is usual, a small space all around for the circular ligament, by which it was retained in place.

Upon the tympanic side of this *basis*, and almost filling up the *fossa*, there was a quantity of membranous substance, which connected itself with the cellular structure already described. The *fenestra rotunda* was covered by the same.

Muscles.—The muscle of the malleus *(tensor tympani)* was strongly developed: its tendon was short and very thick, and its attachment peculiar, as above mentioned. The muscle of the stapes *(stapedius)* existed, but there was no tendon at the small orifice at the summit of the pyramid. Thus, in regard to both these muscles, the development seemed in accordance with the function required of them. In the one case, as no stapes existed, the *stapedius* was only partially developed. In the other, the tensor tympani had alone to keep the membrana tympani in place, and it seemed to do so by its short thick tendon.

The other parts of the organ appeared to be in a normal state.

Left Ear.—External ear and meatus were healthy. On cutting away the meatus, the membrana tympani was found to have been entirely destroyed, with the exception of a very thin slip at the anterior inferior edge. The tympanum was thus fully exposed: it contained a quantity of yellowish, fetid matter, and its lining membrane was completely disorganised.

Ossicula.—The *malleus*, though its *handle* was depressed, was still attached to the incus; but the union was slight, the ligamentous and mucofibrous connections being involved in the general disorganization.

The *incus* was in place: its long *crus* extended to the stapes, but it merely rested upon the latter, the bond of union being broken up, and the least motion separating the bones.

The *stapes* seemed to stand merely in its *fossa* over the fenestra ovalis; it was not bound down by its appropriate band, being easily moved from its position. The ligament *(ligamentum annulare bas. stap.)* forming the important connection between its base and the surrounding bone was entirely destroyed.

The tendon of the *tensor tympani* was disorganised, and that of the *stapedius* destroyed.

The whole condition of the tympanum showed a recent active suppuration, which did not confine itself to this part: the vestibule was penetrated, as well as the mastoid cells; the latter were covered with pus.

The cochlea and one of the semi-circular canals were examined a day or two subsequently—but there was nothing remarkable about them, at this time, worthy of note.

Having completed the examination, it became a matter of great interest to me to know something more of the man's hearing, and in reply to my inquiries on the subject, Dr. Cunningham was good enough to address me a letter in which he says, "The man had resided in the house six months, and it was observed by the nurses of the hospital that his deafness had considerably increased from the time of his admission to that of his death. During the latter part of this time, the difficulty of hearing was so great as to preclude any conversation with him." "An inmate of the house," continues Dr. C., "has lately informed me that he knew W. some six or seven years ago, at which time he was in the habit of attending the markets: he was somewhat deaf even at that time, but could hear when spoken to in a moderately loud tone. He also informed me that he noticed W. when addressed, to incline his *left* ear to the speaker, from which I suppose, we must infer, that the loss of the faculty was less on that side."

From this information, together with the facts disclosed by the *post-mortem* investigation, I think we are justified in the conclusion, that the faculty of hearing in the right ear was either entirely wanting, or was very much impaired ; for the custom of turning the head round, so as to make use of the left ear, must have been very decided to have been remembered after the lapse of years ; it was doubtless a habit that had become identified with his person. Independently of the general inference that every deviation from the normal state of an organ must be attended with an impairment, if not a loss of the function of that organ, the irregularity in this (right) ear was very great, and obviously calculated from the known laws for the propagation of sound, to impair its communication.

Had the membrana tympani been capable, in its abnormal state, of communicating sonorous vibrations, the relation of the malleus, in contact with it, to the parts within, would have conducted them indirectly to the nervous expansion within the labyrinth ; for it will be recollected, the chain of ossicles was broken up by the absence of an important link. But the tense condition of the membrana tympani, as it here existed, was not well calculated for the communication of sound, had the other parts of the organ been perfect. The experiments of Savart and Müller show that under this state of a membrane, the intensity in the communication of sonorous undulations is much diminished, and Dr. Wollaston, in applying the principle to the membrane of the ear, found by experiment, that a temporary imperfection of hearing always resulted when it was made tense. It is true that the experiments of Dr. W. showed that this incapacity had reference only to certain sounds ; (for when the membrana tympani was made tense, grave sounds only were rendered inaudible ;) still, for the perfection of hearing conversation, there should be an equal capability both for acute and grave sounds.

As to the influence upon the function of the ear, of the several other parts in their abnormal condition, many suggestions offer themselves to the mind. It may be worthy a passing inquiry, how far nature may have intended, by bringing down the handle of the malleus in such close proximity to the walls of the labyrinth, to compensate for the usual communication, through the chain of bones, to the vestibule.

One more remark may be made; that if the membrana tympani had been capable of communicating vibrations to the *air* within the tympanum, the base of the stapes, or operculum, like the same moveable plate in some of the reptiles without a tympanum, offered no impediment to the communication of the undulations to the fluid of the labyrinth.

In regard to the condition of the *left ear*, the result of disease, we are much more familiar, and can readily understand, under the supposition that the right ear was unfit to perform its function, how the hearing of the individual, towards the close of his life, became so much impaired, or was probably entirely lost. It is likely, judging from the state in which it was found *post-mortem*, that the membrana tympani was for a long time in a state of ulceration, with perforation; and that this was its condition six or seven years ago, the time spoken of in reference to the hearing. This is a very common cause of impaired hearing in scrophulous persons. My observations in numerous cases have shown that in such cases, where the perforation is of small extent, the hearing is only partially lost or slightly impaired. Where the ulceration extends, so as to produce entire loss of the membrana tympani even of both ears, although the hearing is much more impaired, yet if the disease be limited to this membrane only, it is not so much injured as would be, *à priori*, supposed. Even when the disease is allowed to extend to the parts within the tympanum, and the *malleus* and *incus* come away, if the ulceration do not embrace the ligament and membranes binding the *stapes* to its place, the loss of the faculty is still not great. Should, however, this part become affected, the stapes generally comes away, the inner structure of the ear becomes involved, and loss of the faculty is the result.

I cannot but believe, that such was the progress of disease in this ear. The increase of the deafness was doubtless owing to the increase of the ulcerative process in the ear, corresponding to the development of the same condition in the lungs. The whole membrana tympani was destroyed; the fibrous or fibro-mucous tissue of the tympanum became implicated; the ligament or membrane connecting the stapes to the fenestra ovalis was destroyed, and the inner and more important parts of the organ were involved. That this extension of the ulceration could not long have existed, to the degree found at the post-mortem examination, is probable, from the position of the ossicles. I have generally found that when this destructive disease is not soon arrested, the malleus and incus come away. In the present case, the falling off of the vital powers seems to have involved the whole texture of the tympanum at once; the bones were undisturbed in their position, the stapes stood in its place, whilst all the surrounding textures were destroyed, and very great, if not entire loss of the faculty was the result, in the short period he remained in the hospital.

Instances of irregular development of the ossicula auditûs, and other malformations of the ear, are not entirely wanting in the works of anatomists. Pathologists, however, have had few opportunities of presenting such cases in connection with impaired function of the ear, previously observed. Morgagni* makes mention of two cases, certain of the features of which bear some resemblance to our present one. In speaking of the various causes of deafness, he says, "It is not to be doubted but that their number may be increased every day, as for instance, when *innumerable membranes drawn from all sides, and intersecting each other, mutually occupy the whole cavity of the tympanum;* as I found in that ear, in regard to which marks were not wanting, that the man had not heard at all, or at least very little; and in like manner, *when any muscle out of those which serve to the motion of the little bones in the tympanum is immovable and contracted*, as I found in another man, who, when he was alive if any one could have divined, would not, I believe, have been interrogated to no purpose at all, concerning the certain disorders of that ear."

It may be well to observe before concluding, that if the abnormal state of the incus and stapes, as found in the present case, be the result of an arrest of development, it would go to confirm the opinions of Meckel rather than those of Cassebohm in regard to the order of development of the *ossicula:* for the details of which I beg leave to refer to the well known work on anatomy of the former. And in regard to the small portion of the stapes that exists in our case, the remarks† of Professor Tiedemann of Heidelberg seem apposite. In describing two cases of peculiarly formed *stapedes*, he remarks that such is the infinite variety presented by the animal organization in all its parts, that we cannot fail to observe a well marked gradation from the most simple to the most perfect of beings, and that every aberration in the form of any of the organs of the upper groups approximates them to that of the inferior ones. In illustration of which, Professor T. describes two cases, one of them in a new-born child, the other in an adult, where the *stapedes* were so altered in form as closely to resemble the *columella* of the *ornithorynchus*, the *echidnas,* and that of birds and reptiles.

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* De Sedibus et Causis Morborum.

† Journal Complem. du Dict. des Sc. Med. Nov. 1820.

In the case described in this paper, does not the absence of every part of the *stapes,* with the exception of the *base,* liken it to the osseous operculum found in the *bombinatores, land salamander* and *cæciliæ ;* of the effect of which, in the communication of sonorous undulations I have already spoken. ?